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Sarcoidosis Presenting as a Proximal Phalangeal Bony Swelling—a Case and Review of the Literature

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Abstract We report a case of a 27-year-old man who presented with a progressive painful swelling at the base of his left index finger, with radiographs and a computed tomography scan revealing a lytic lesion of the proximal phalanx. Following further investigation, the patient underwent a bone biopsy that revealed a florid noncaseating granulomatous chronic inflammatory infiltrate, compatible with sarcoidosis. Osseous sarcoidosis of the hand is uncommon and, in the absence of significant systemic disease, is rarely the primary presenting feature. Early diagnosis and treatment of such undetermined bone

pathology, via referral to a regional musculoskeletal tumor service, can prevent significant future complications.

Keywords Caseating granuloma · Osseous sarcoidosis · Phalanx · Sarcoidosis

Introduction

Sarcoidosis is a systemic disorder of undetermined etiology that is characterized by the deposition of noncaseating granulomata, most commonly in the lungs [13]. Diagnosis may be difficult due to the wide variety of potential presentations.

Published literature reveals a wide range in the incidence of sarcoidosis involving bone (1–30%) [8, 21]. Sarcoidosis of the hand is said to occur in 0.2% of cases and, in the absence of significant systemic disease, is rarely the primary presenting feature [4, 9, 15, 21].

We present the case of a young patient who presented with left index finger pain and swelling due to sarcoid of the proximal phalanx. He exhibited no additional clinical manifestations of the disorder. Imaging showed mediastinal and generalized lymphadenopathy, splenomegaly, and normal lung parenchyma.

Case Report

A 27-year-old man presented to his general practitioner with a 1-month history of a progressive painful swelling over the proximal phalanx of his left index finger. He was referred to the orthopedic department of a district general hospital. There was no history of trauma. He had no respiratory symptoms, weight loss, or night sweats. Exam-

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ination revealed a bone-hard swollen proximal phalanx with overlying cutaneous erythema without significant heat and with a good range of movement. The rest of the examination was unremarkable. There was no significant past medical history. His grandfather, with whom he had had contact in the past, had pulmonary tuberculosis several decades before.

A radiograph of the finger (Fig. 1) showed an ill-defined lytic lesion of the proximal phalanx with cortical thinning and adjacent soft tissue swelling. There was no lesional calcification. Full blood count revealed a lymphopenia ($1.06 \times 10^9/l$, normal range $1.5\text{--}4.0 \times 10^9/l$), but was otherwise normal. Urea and electrolytes, liver function tests, calcium levels, C-reactive protein, and erythrocyte sedimentation rate were all normal. A myeloma screen was negative.

A computed tomography (CT) scan of the affected finger (Fig. 2) showed an irregularly margined lytic lesion causing endosteal thinning of the cortex of the proximal phalanx with overlying soft tissue swelling. There was no calcification. A radionuclide bone scan (Fig. 3) confirmed increased activity in the digital lesion. The remainder of the skeleton was normal.

The patient was referred to the regional musculoskeletal tumor service where open bone biopsy and curettage was



Figure 1 AP and lateral radiographs of the left index finger. There is a poorly defined lytic lesion in the diaphysis and distal metaphysis of the proximal phalanx. A ground-glass matrix predominates and there is thinning of the cortex. No foci of calcification or periosteal reaction are present and there is minor soft tissue swelling.

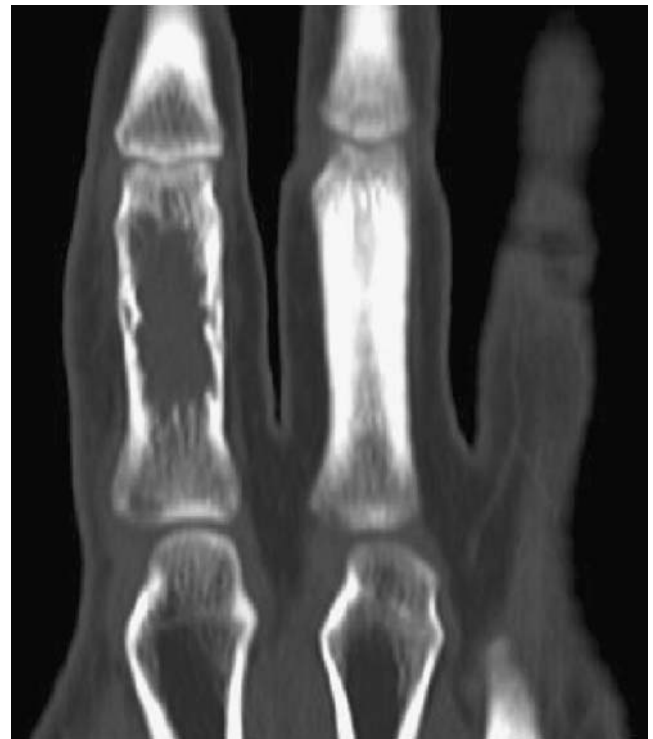


Figure 2 Coronal CT of the left index finger. The lytic lesion in the shaft of the proximal phalanx lesion causes cortical thinning. The endosteal margin is irregular and linear tracts extend into the thinned cortex. There is no periosteal reaction or calcification.

undertaken under general anesthesia. Histological examination revealed a florid granulomatous chronic inflammatory infiltrate destroying the bone of the proximal phalanx. The granulomata were small and noncaseating (Fig. 4). Ziel-Neelsen and Wade-Fite stained sections showed no evidence of acid and alcohol fast bacilli. No organisms were seen on microscopy or grown on routine or tuberculosis culture. There was no evidence of neoplasia. Subsequent chest radiograph (Fig. 5) showed bilateral symmetrical hilar enlargement. A CT scan of the chest (Fig. 6) showed bilateral hilar subcarinal, aortopulmonary, and paratracheal lymphadenopathy. Lung parenchymas were normal but minor pleural beading was present. Bilateral axillary, epicardial, porta hepatis, and celiac axis lymphadenopathy and splenomegaly were also present. Angiotensin-converting enzyme level was mildly elevated (69 U/l , normal $16\text{--}63 \text{ U/l}$) on subsequent blood tests. The clinical, laboratory, imaging, and histological findings were considered to be consistent with a diagnosis of sarcoidosis. The patient was treated with local intra-osseous corticosteroid injection.

At 1 year follow-up, the patient is currently on a reducing regime of oral corticosteroid. His left index finger has no significant functional deficit with only postsurgical changes noted. No respiratory symptoms are present and a



Figure 3 Radionuclide bone scan. There is increased uptake in the left second finger (*arrows*). The scan is otherwise normal.

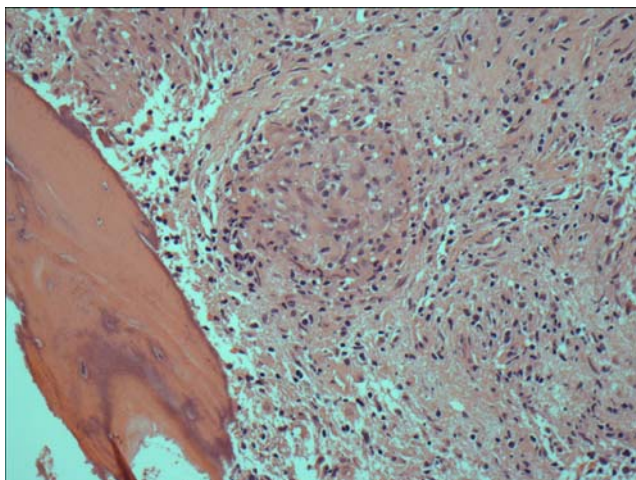


Figure 4 A hematoxylin and eosin stained section showing tight noncaseating granulomata within the medullary space (original magnification $\times 400$).



Figure 5 PA chest radiograph showing bilateral hilar lymphadenopathy.

repeat chest radiograph revealed that the bilateral hilar lymphadenopathy had significantly improved.

Discussion

Sarcoidosis is a rare idiopathic multisystem disease that is characterized by the deposition of noncaseating epithelioid granulomata [13]. The most frequently affected sites are the lungs and lymph nodes, but the liver, heart, skin, eyes, and musculoskeletal, endocrine, and nervous systems may also be involved [13]. Rizzato suggested that more than 30% of patients with sarcoidosis have extrapulmonary manifestations at initial presentation [18].

Osseous sarcoidosis is uncommon, often asymptomatic, and is usually associated with skin lesions [5, 13, 21]. Soft tissue involvement is more frequent [6, 14]. Cutaneous lesions without bone involvement affect approximately



Figure 6 Axial contrast-enhanced CT scan of the thorax showing subcarinal and bilateral hilar lymphadenopathy.

25% of sarcoid patients [11, 13]. The hand is the most common skeletal site of sarcoidosis [24], but is seldom the primary presenting feature and is usually associated with progressive pulmonary manifestations of the disease [15, 21]. The middle and distal phalanges are more frequently involved than the proximal phalanges or metacarpals [22]. The characteristic radiographic features are replacement of the normal trabeculae by a honeycomb or latticework pattern [16, 17]. Lytic lesions may cause endosteal scalloping, as in our case, and may be focal, central, or eccentric, or involve the whole bone [16, 17]. Periosteal reaction is uncommon. Bone expansion and fractures, soft tissue swelling, sclerosis, and acroosteolysis have been reported [1, 4, 15, 20, 21]. With digital involvement, rupture of the extensor tendon of the finger may occur [20].

Bone biopsy is key to a definitive and early diagnosis [12, 23] because the clinical and radiological features are often nonspecific. It is essential to rule out other potential causes for lytic bone lesion such as primary and secondary bone tumors and nontumorous conditions. In the case of granulomatous disease, typical or atypical mycobacterial infection should be excluded before definitive treatment is commenced.

The treatment of extrapulmonary sarcoidosis remains controversial. Oral corticosteroids are recommended for osseous sarcoidosis, although they are not curative, and there are conflicting opinions on their long-term efficacy [3, 13]. In this case, we decided to initially use local corticosteroid injection, with subsequent employment of oral corticosteroid. Antimalarial drugs, such as hydroxychloroquine, have also been prescribed effectively [2, 19].

Open curettage for symptomatic lesions appears to be the most commonly employed primary surgical treatment for osseous sarcoidosis affecting the hand [4]. Generally, good results have been seen within the limited published data [4, 9, 10, 15]. One paper reported the use of repeated curettage with excision and iliac bone grafting for recurrence post primary surgical treatment [4]. The aggressive and recurring nature of the osseous sarcoidosis in this case led to resorption of the bone graft. Amputation may be necessary for severe and unsalvageable finger disability [10]. Concurrent use of corticosteroid therapy is often employed, although initiation is patient dependant [3, 4, 13, 21]. Postmedical and/or surgical treatment, regular long-term follow-up is recommended for all patients [7, 13].

Sarcoidosis provides diagnostic, therapeutic, and prognostic challenges. A wide range of systemic presentations is possible and early diagnosis and treatment may prevent potentially significant complications. Osseous sarcoidosis of the hand is rarely the primary presenting feature in the absence of clinically significant systemic disease. Where bony disease is identified, staging investigations must

include a radionuclide bone scan to confirm the extent of skeletal involvement. Treatment of systemic disease should parallel local bony surgery or injection to provide the most comprehensive chance of cure.

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